

and nodular. They resemble metastatic lesions and predominate in the peripheral portions of the lower lobes, chiefly in young males. Frequently there is an accompanying cutaneous angitis and panniculitis. Nodular renal lesions resembling those in the lungs can occur, but there is not the glomerulonephritis of the Wegener triad. The central nervous system is involved in one-fifth of the fatal cases. The condition occasionally terminates in atypical lymphoma. Pulmonary lesions may be asymptomatic or associated with fever and non-productive cough. Treatment with steroids may be associated with remission or arrest of the process, but this outcome can occur spontaneously.

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#### REFERENCE

Liebow AA, Carrington CB: Lymphomatoid variant of "limited Wegener's granulomatosis" (Abstract Section). *Am J Pathol* 55:78-A, Jun 1969

### Pulmonary Veno-Obstructive Disease

Intrapulmonary veins can be specifically occluded by thrombosis and organization. The first manifestation may resemble influenza, but more commonly the onset is insidious, with increasing dyspnea as the chief complaint. One-third of the patients are less than 16 years of age. There is progressive right-sided heart failure without evidence of left atrial enlargement. Radiographically wandering pulmonary infiltrates are seen and Kerley lines are prominent. Angiography may reveal focally delayed emptying of pulmonary arteries. Pulmonary wedge pressure may be elevated or normal. The cause is unknown and no method of treatment is available.

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Heath D, Scott O, Lynch J: Pulmonary veno-occlusive disease. *Thorax* 26:663, Nov 1971

### Microangiopathic Hemolytic Anemia

The sequential activation of the coagulation factors terminating in the production of thrombin and the formation of a fibrin clot normally occurs in the interstices of a platelet plug. On occasion, due to disease, the activation of the procoagulant enzymes occurs widely in the bloodstream and disseminated intravascular coagulation (DIC) results. Whereas DIC is always a secondary mechanism of disease, microangiopathic hemolytic anemia (MAHA) is tertiary. It results from DIC of a certain type and duration.

Schistocytes, red cell fragments of characteristic morphologic pattern, are the hallmark of MAHA. Fibrin microclots produced by DIC are filtered out of the circulation in arterioles, primarily those of lung and kidney. Rapidly moving red cells are sheared by these fine fibrin strands the way cheese can be cut by a taut wire. The resulting damage decreases the ratio of membrane to hemoglobin and hence the deformability of the schistocytes. The most severely damaged cells lyse, releasing hemoglobin. Less severely traumatized cells circulate briefly and are removed within a few hours by the spleen. Only those cells which still possess a high enough ratio of membrane to hemoglobin are deformable enough to persist in the circulation as schistocytes.

In addition to the obvious sequel of anemia, MAHA causes hemoglobinemia as a result of intravascular lysis of severely damaged cells; bilirubinemia due to increased splenic destruction; thrombocytopenia due to DIC; and, should the process become subacute, erythroblastemia as the bone marrow releases nucleated red cells into the peripheral circulation in an attempt to compensate for hemolysis.

Postmortem tissues of patients who had MAHA during life often do not show the microclots of fine fibrin strands that cause the red cell damage, for they are rapidly incorporated into coarser, more amorphous deposits of fibrin. Strongly